Surgical Treatment of Infantile Hydrocephalus

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• The operation of endoscopic coagulation of the choroid plexuses for the relief of infantile hydrocephalus is now 18 years old. Nearly a hundred cases have been reported and the indications and procedure are well standardized. Several patients have grown up apparently normally from the earliest series of operations.

In a recent series of 20 operations performed on ten patients in the past ten years, there have been no deaths attributable to the procedure. The operation has substantially decreased the pressure in all cases, and has brought it within normal limits in all cases in which it was performed before the head became grossly enlarged.

The mentality has improved following relief of pressure in all cases. When the operation was performed before deterioration began, the results were uniformly excellent.

THE CONDITION of infantile hydrocephalus is unmistakably portrayed in ancient Greek votive statuettes, but the first written description of it apparently was that of Celsus.[1] He described a disorder of infants "where the fluid distends the skin, causes a swelling of it, and yields to the pressure of the finger: This the Greeks call hydrocephalus."

This early description calls attention at once to the fundamental characteristic of infantile hydrocephalus—the increase of intracranial pressure. The remaining clinical details were gradually filled in during the 19th century; namely, that hydrocephalus is a common disorder, occasionally present at birth but more likely to come on during the first year of life; that it is often associated with spina bifida, and may be precipitated by the repair of a meningocele; and that children afflicted with the condition sometimes survive to adult life, occasionally with intelligence unimpaired. Unfortunately, a serious error was introduced by the pathologic anatomists of that period—confusion between the condition of increased intraventricular pressure or true hydrocephalus according to Celsus, and the enlargement of the cerebral ventricles resulting from atrophy, formerly called "hydrocephalus ex vacuo."

The distinction between spontaneous infantile hydrocephalus and tuberculous meningitis became clear with the introduction of lumbar puncture. Recognition of the physiologic features of the condition resulted from the work of Dandy and Blackfan in 1918 and laid the foundation for a sound surgical approach. Gradually during the following two decades the condition of subdural hygroma became familiar to pediatricians, and the surgical treatment became established. Cases of hydrocephalus resulting from obstruction of the aqueduct by tumors, atresia or inflammatory diseases (such as toxoplasmosis) have been reported from time to time.

This presentation deals solely with the treatment of spontaneous communicating infantile hydrocephalus, with or without meningocele. Medical treatment of this condition appears now to be abandoned and spontaneous recovery is so rare as to elude statistical evaluation. A variety of surgical procedures are now available, however. One of them, to be described in detail, has been well standardized for 18 years and can now be performed with little hazard and with a high proportion of excellent results in well selected cases.

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DESTRUCTION OF THE CHOROID PLEXUSES

The conception that the spinal fluid is secreted by the choroid plexus originated with Magendie, and isolated attempts (on the whole unsuccessful) to relieve hydrocephalus by destruction of the plexus were carried out in the early years of this century. The possibilities of surgical treatment were, however, first systematically explored by Dandy between 1918 and 1922.[2, 3] He showed experimentally that removal of the plexus caused collapse of a blocked lateral ventricle in animals, and described a technique for removal of the plexus under direct observation in cases of non-obstructive hydrocephalus.[2] He did not present statistical studies of his cases. In 1932[4] he stated that "the survival period has not been long enough to be certain of cures," and in 1938[5] "I have had several undoubted cures resulting from this method."

The author's technique was published in 1934.[6] It is based on the use of a distinctive type of coagulating endoscope with a rod of optical glass serving as a viewing system and also as a carrier for two electrodes and a lamp. The instrument gives several

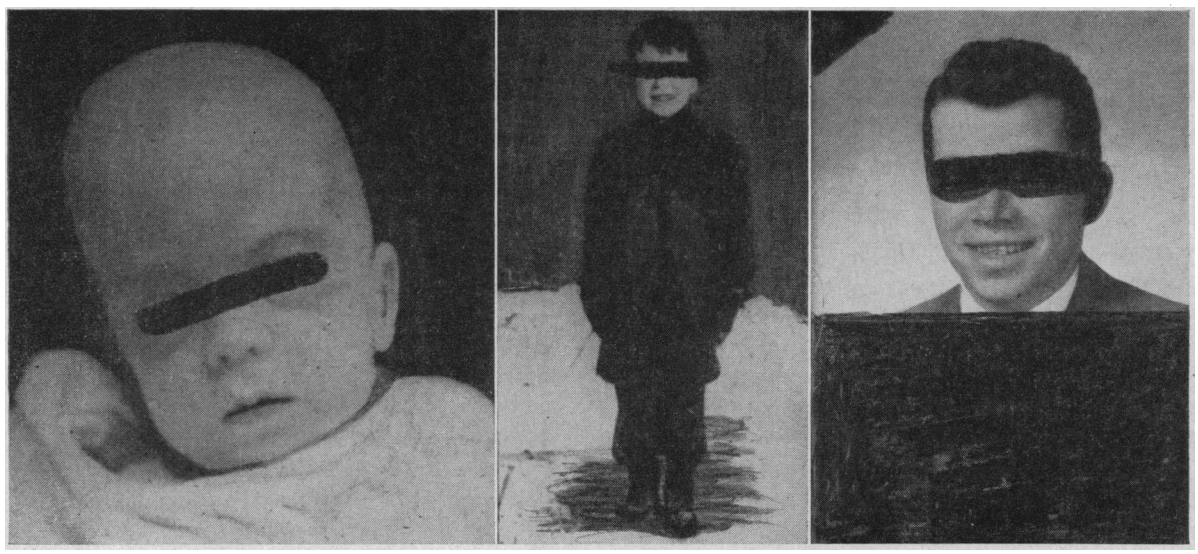

Figure 1.—*Left,* photograph of patient in third month of life just before coagulation of choroid plexuses. *Center,* patient at six years of age. *Right,* at time of graduation from high school.

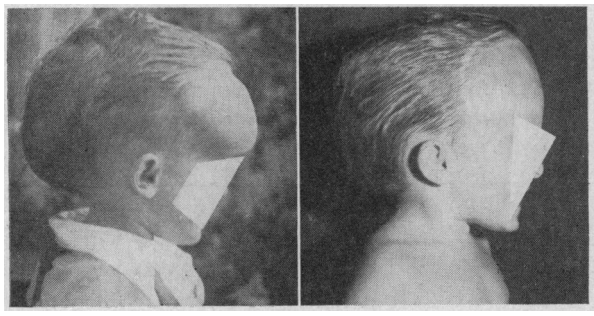

Figure 2.—*Left,* patient at three years of age just before endoscopic coagulation of the choroid plexuses. *Right,* two and one-half years later, after reduction of intracranial pressure had permitted plastic repair of elevated flap of transfrontal operation performed elsewhere. Intracranial pressure was normal at last report.

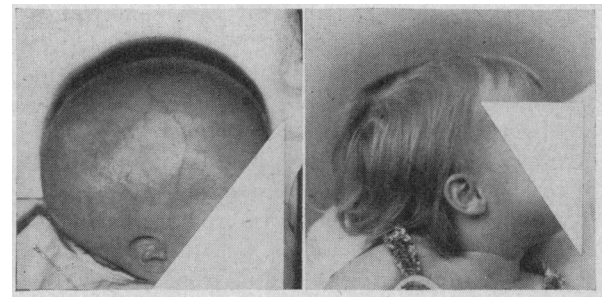

Figure 3.—*Left,* patient at five months of age just before coagulation of choroid plexus. *Right,* patient a little more than two years later. At time of last report, intracranial pressure was normal.

advantages. First, it provides an extremely wide optical aperture, which permits visualization of the ventricular contents even when the fluid is bloody. Second, no current passes through the brain; there is a small intense electrical field between the electrodes. Third, the diameter of the instrument may be kept sufficiently small to permit introducing it into the frontal and temporal horns even before the ventricles have become greatly dilated. This instrument was developed after unsuccessful trial of conventional endoscopes bearing unipolar coagulating electrodes. The employment of these tended to produce necrosis of the floor of the ventricle, and the operative field was poorly seen. An endoscope of the conventional type, with provision for irrigation, was devised by Scarff in 1936[8] and he still employs it, with excellent results.[9, 10] In the most recent series of 19 cases reported upon by Scarff there was only one "operative" death, and the result was excellent in 15 cases. Other surgeons have reverted to Dandy's method, but no significant statistics are reported.

RESULTS

The author's results were last fully reported in 1943.[7] Disregarding other conditions, 71 operations were reported on 42 patients, with 11 operative deaths, of which seven occurred in the first 17 cases. Since then, 20 operations of this type have been performed, upon ten patients, with no operative deaths.

It has not been possible to get continuing follow-up reports on three of the patients, who were operated upon in New York between 1943 and 1946, but all were doing well in 1946. Of the seven patients operated upon since 1947, four are still alive and doing well, with maintenance or improvement of mentality. One patient who was incompletely re-

lieved by cauterization of the choroid plexuses and then by an additional ventriculomastoidostomy, has done well since a tube was inserted from the subarachnoid space into the peritoneal cavity. One was apparently relieved of hydrocephalus but died of unrelated causes. One died following an attempt at third ventriculostomy.

Since many detailed case reports have been presented by Scarff[9, 10] and by the author in previous publications, only a few general observations on the indications for operation, on details of technique and on results need be set down here.

First, this operation is a safe one—safer perhaps than ventriculography in such cases, and apparently safer than the recently proposed drainage operations. In the early stages of the disease it is probably safer to operate than to wait in the hope that it will become arrested spontaneously.

Second, the outlook is excellent if patients are referred for operation before the mentality is affected and before there is irreparable damage to the brain. Ideally, the decision for operation should be made when the presenting symptom is a swelling of the fontanelles and infantile hydrocephalus has been diagnosed on the basis of a ventricular puncture with the patient under sedation—a procedure which serves to rule out meningitis, toxoplasmosis, and subdural hygroma and usually renders ventriculography unnecessary.

Third, prognosis depends chiefly upon the patient's intellectual endowment at the time of operation, but the degree of enlargement of the head and thinning of the cortex are limiting factors. Sometimes a patient with enormous ventricular dilation may have an unexpected degree of intelligence and Scarff[9] has shown that the ventricular wall may grow thicker following relief of intracranial pressure. Gross enlargement of the head may produce secondary adhesions about the base, in which case destruction of the plexuses may not completely relieve the pressure. Many of the poor results, both in the author's series and in Scarff's, would undoubtedly have been avoided had the patients been operated upon sooner.

Fourth, operation may be considered justified even when the patient has mediocre mentality and gross enlargement of the head. Such persons do not necessarily die; they sometimes grow up to adult life with monstrous heads and progressively impaired mentality. It may seem wiser to attempt to arrest the accumulation of fluid and preserve the remaining mentality. Despite relief of pressure, however, patients with poor mentality appear to have a poor life expectancy.

Fifth, ventriculostomy of various types may prove helpful as an adjuvant to cauterization of the plexus, which appears to be the operation of choice in cases of communicating hydrocephalus. Certainly the drainage operations are more likely to succeed and less likely to lead to complications if the production of fluid is restricted.

Sixth, certain technical refinements have been added to the technique published by the author in 1943.[7] Suture of the pia to the dura minimizes the danger of collapse of the ventricle at operation, or the formation of subdural cysts. Suture of the periosteum over the suture line decreases the danger of leakage. Drainage of the ventricle through a tiny plastic tube diminishes the postoperative reaction (and in one case this device made it possible to operate on both sides at one time).

Seventh, always after cauterization of both sides, reinspection should be carried out to make quite certain that no tags of plexus were left behind. This is particularly important if the preliminary operations were performed with a ventriculoscope of the cystoscope type.

Eighth, coagulation of the plexuses may be of great help in the treatment of other conditions involving increase of intracranial pressure—for example, postmeningitic hydrocephalus, and large, tense, sessile meningoceles which do not destroy the function of the sphincters.

DISCUSSION

Infantile hydrocephalus is a common condition, perhaps as common as harelip or clubfoot. The condition is easy to recognize, even in early stages. There is a well established surgical operation which entails little hazard and offers an excellent prospect of relief in early cases. To permit one of the unfortunate children with promising mentality to slip past the period of election without a trial of operation is no more conscionable than to permit a patient with meningitis to go without antibiotic drugs.

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ADDENDUM

After this communication was written, a brief review of a presentation of the surgical treatment of hydrocephalus by Walker* became available. It is summarized in the following quotation:

On the basis of the medical literature and the records of the Johns Hopkins Hospital, the results at the end of two years were as shown in the tabulation:

Operation	No. of Cases	Operative Mortality	Results After Two Yr.† Alive	Dead
Plexotomy	194	70	31	105
Third ventriculostomy	197	44	39	81
Arachnoido-peritoneostomy‡	105	12	41	6
Arachnoido-ureterostomy	86	6	17	28
Ventriculo-cisternostomy	25	8	9	9

† Only the patients whose states were definitely known two years after operation coded.

‡ Based largely on the series of Arendt whose results are not well documented.

It is not clear whether "plexotomy" includes endoscopic coagulation, or whether it refers to Dandy's operation of removal of the plexus. In either case, a comparison between this table and the results reported in the present article speaks for itself.

* Walker, A. E.: A Critique of Surgery for Hydrocephalus (abstract of society presentation), Archives of Neurology and Psychiatry, 67:822-823, June 1952.

New Film Catalog

A REVISED LIST of medical and health films has been prepared by the A.M.A.'s Committee on Medical Motion Pictures. Brief descriptions, running time, and rules and regulations are included in the catalog for 78 medical films which are available from the committee. Copies of the list may be obtained from the committee.—*From A.M.A. News Notes.*